Asymptomatic Wolff-Parkinson-White Syndrome in Children

An Unnatural History?*

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The management of an asymptomatic child incidentally discovered to have Wolff-Parkinson-White (WPW) syndrome on electrocardiogram (ECG) is a controversial issue. One big impediment to settling the controversy is our ignorance about the natural history of asymptomatic WPW syndrome. In this issue of the Journal, Santinelli et al. (1) report the largest cohort (to date) of children with asymptomatic WPW syndrome intensively followed for a period of time. Patients were identified incidentally or through the mass ECG screening program for athletes instituted in Italy since 1982.

All children (age 8 to 12 years) underwent an electrophysiology (EP) study at the start of the prospective protocol. They were followed every 6 months with an ECG and a 24-h Holter monitor. The primary end point was the first arrhythmic event. Over a median follow-up of 57 months, 51 of 133 children became symptomatic. Of these, 19 had potentially life-threatening arrhythmias. Interestingly, some children with potentially life-threatening arrhythmias had minimal (n = 5) or atypical symptoms (n = 8) such as nausea, sudden tiredness with anxiety, abdominal pain and swelling, and inability to concentrate while playing. Factors associated with increased risk were the presence of multiple accessory pathways and an accessory pathway effective refractory period (APERP) <240 ms. Atrioventricular re-entry tachycardia (AVRT) inducibility did not prove significant on multivariate analysis. What are the lessons of this study and how do we incorporate its findings into clinical practice? Some background first . . .

While it commonly presents with paroxysmal AVRT, WPW syndrome may rarely cause sudden death (SD) due to atrial fibrillation (AF) being rapidly conducted to the ventricle over the accessory pathway resulting in ventricular fibrillation (2,3). In some patients, SD may be the presenting symptom. Deal et al. (4) reported 42 SDs in children with WPW syndrome. Of these, in 20, SD was the presenting symptom.

The approach to the asymptomatic patient is now significantly impacted by the availability of EP and catheter ablation (CA). However, EP and CA are invasive procedures with a potential for complications that could be unacceptable for the asymptomatic patient; hence, the need for careful and objective risk-benefit analysis.

EP can help identify patients at risk for developing symptoms and SD due to WPW syndrome. Inducibility of AVRT, a shorter APERP (<240 ms), a shorter pre-excited RR interval during induced AF (<240 ms), the presence of multiple pathways, and septal and right-sided pathway locations appear to identify a higher-risk group (3,5–7).

Some authorities have suggested that noninvasive risk stratification with Holter monitoring, exercise stress testing, and pharmacologic testing should be performed before invasive studies are considered (8). However, the sensitivity and specificity of noninvasive testing has been shown to be poor (9,10).

Transesophageal EP may be a semi-invasive technique to avoid the potential vascular complications of intracardiac EP (11). However, transesophageal EP is not entirely risk free. High output pacing may be required to activate the atrium from the esophagus, which can be painful and requires the use of heavy sedation (12).

The group from Milan has recently made major contributions to our understanding of the asymptomatic patient with WPW syndrome. They reported the ability of EP testing to risk stratify asymptomatic patients (13). Of the 162 patients (adults and children) studied, only 4 of the 115 patients noninducible for supraventricular tachycardia (SVT) went on to develop clinical SVT during a mean follow-up of 37.7 months. Of the 47 inducible patients, 21 developed SVT and 8 AF. And, of these 8 with AF, 2 had near-miss SD, and 1 died suddenly.

The same group also showed that prophylactic CA in asymptomatic WPW syndrome patients (children and adults) reduces the likelihood of arrhythmia (14). Of 37 patients with inducible SVT (at EP study) who underwent ablation, only 2 had SVT at follow-up (both had AV nodal re-entry tachycardia) while of 35 with inducible SVT randomized to no treatment, 21 had SVT at follow-up.

Their previous study in children between 5 and 12 years of age also suggested that prophylactic CA significantly reduces symptoms (15). Of those with inducible SVT, only 1 of the 27 randomized to ablation developed SVT whereas 7 of 20 randomized to no treatment did, and 1 from the latter group died suddenly.

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The major message of the studies from Milan would be that the natural history of asymptomatic WPW syndrome in children is not benign and that EP study provides excellent risk stratification. Patients who are noninducible with a long APERP may be followed without treatment, but those who are inducible or have a short APERP should be considered for ablation. This take-home message is at variance with previous studies and with current guidelines from major cardiology organizations.

Prior studies suggest that SD is rare in an asymptomatic patient with WPW syndrome. Leitch et al. (16) showed no SDs in 75 initially asymptomatic patients followed for a median of 4.3 years. The population-based study of the natural history of WPW syndrome by Munger et al. (17) had 113 WPW patients of whom 53 were asymptomatic at detection. While there were 2 SDs in the symptomatic group, there were none in the asymptomatic patients. Goudevenos et al. (18) followed 157 patients with incidentally discovered WPW syndrome for an average of 55 months with no SDs in their cohort. Inoue et al. (19) followed 57 children detected to have WPW syndrome during screening ECGs for almost 8 years on average, again with no SDs.

Current guidelines from the American Heart Association, American College of Cardiology, and the European Society of Cardiology restrict ablation for asymptomatic WPW syndrome to those in high-risk occupations and professional athletes (20). The Heart Rhythm Society expert consensus is that ablation for asymptomatic WPW syndrome in children age >5 years is a class IIB indication and that in those age <5 years it is a class III indication (21).

The main difference between prior studies and the study of Santinelli et al. (1) is the intensity of monitoring and the credence given to atypical symptoms, which are often dismissed as being noncardiac, in the current study. This study, therefore, alerts us to the importance of these atypical symptoms in WPW patients.

Interestingly, current practice by pediatric electrophysiologists may be more in line with the Milan group recommendations. Campbell et al. (22) surveyed the members of the PACES (Pediatric and Congenital Electrophysiology Society) study group. Of 43 respondents (of whom 37 had been performing ablation for >5 years), 36 used EP study to risk-stratify children with asymptomatic WPW syndrome. Most (33 of 43, 77%) would also perform ablation on children with a shortest pre-excited RR during AF of <240 ms, and 19 of 43 (44%) would have ablated those with APERP <240 ms. Only 11 of 43 (26%) would ablate those with inducible SVT alone. Special consideration of ablation was given by these physicians to patients considering high-risk careers; competitive athletes; and those with coexisting congenital heart disease, asthma, and attention deficit disorder likely to require medical management with drugs.

There are some caveats to the natural history study by Santinelli et al. (1). First, children <5 years old were excluded by design, and all children were >8 years old. Second, there was a high proportion of competitive athletes picked up during the Italian countrywide ECG screening program for competitive athletes.

In summary, this report by Santinelli et al. (1) further reinforces the notion that children with asymptomatic WPW syndrome should undergo an EP study. However, prudence dictates that noninvasive testing with Holter and exercise test (if the Holter does not show intermittent pre-excitation) should be considered before the EP study. It may also be wise to consider transesophageal EP rather than proceeding directly to intracardiac testing. The exact age at which the EP study should be done is unclear, but >8 years seems a reasonable age cutoff. Those who have a potentially dangerous pathway (as defined by the presence of multiple pathways or a short APERP) should probably undergo ablation as long as it is felt to be reasonably safe. The physician involved needs to use special care and discretion in making this decision and must especially consider the success rate and complication rate in his/her hands. While success rates for CA have been improving, serious complications still occur in a small minority of patients (23,24).

Lastly, the findings of the Milan group suggest a more malignant natural history in their cohort than previous reports (16–19). It is advisable to perform further studies in other ethnic/geographic populations before these findings can be accepted in all countries. Despite these caveats, the findings in this study should prompt a reconsideration of current guidelines by professional organizations in the area of pediatric arrhythmia.

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